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Nasal Septal Chondromyxoid Fibroma: A Case Report & Literature Review

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Abstract

Chondromyxoid fibroma is an uncommon bone tumor and only four documented cases have described it arising in the nasal septum. In the presented case report, an incidentally found mass originating from the ventral septum was suspected to be chondromyxoid fibroma on initial pathology review. Surgical treatment with enucleation, excision of the involved ventral septum, and curettage of the cavity were performed and final pathology confirmed the pre-operative diagnosis. With particular focus on chondromyxoid fibroma of the nasal septum, this literature review highlights incidence and location, patient demographic, symptoms, pathology, surgical treatment, surgical complications, recurrence and follow-up.

Keywords

chondromyxoid fibroma; bone tumor; nasal septum

Introduction

First described by Jaffe et al in 1948, chondromyxoid fibroma (CMF) accounts for less than 1% of all bone tumors [1,2]. Most commonly, CMF presents in the proximal tibia and distal femur, but approximately 2-5.4% arise in the craniofacial bones [2,3]. A mere four cases of CMF presenting in the nasal septum have been recorded in the literature [3]. As such, there is a paucity of knowledge on the surgical management of nasal septal CMF. This is further complicated by difficulty in diagnosing CMF, in general, because it shares many pathologic features with chondrosarcoma [3]. This article aims to describe an additional case of nasal septal CMF in order to bolster the available knowledge on the presentation, diagnosis, surgical treatment, and follow-up of a rare tumor in a rare location.

Case Report

A 71-year-old Caucasian woman was referred for evaluation of a nasal floor mass of identified on a strabismus work-up MRI. The patient reported a several year history of clear rhinorrhea but denied pain, epistaxis, or nasal obstruction. CT and MRI imaging identified a 3.1x3.1x2.8 cm bi-lobed mass (left greater than right, with irregular intralesional enhancement and without internal calcifications) in the bilateral inferior nasal cavities originating from the ventral septum with local mass effect and erosion of the hard palate (see Figure 1). Endoscopic biopsy revealed a low-grade cartilaginous neoplasm with histologic features suggestive of chondromyxoid fibroma.

The mass was enucleated, the ventral septum excised, and curettage of the cavity performed via a midfacial degloving approach (see Figure 2). The final pathology report confirmed the diagnosis of chondromyxoid fibroma. The patient's post-operative course was complicated by left epiphora and was treated using a dacryocystorhinostomy. Follow-up imaging at 4 months did not identify recurrent disease and she was content with the aesthetic outcome.

Discussion

Incidence & Location:

Though CMF primarily presents in the long bones, it can also arise in the craniofacial bones and sinonasal tract [4]. According to a review of 278 cases by Wu et al, 15 of 277 cases presented in the facial/skull bones [4]. Most commonly, the frontal, sphenoid, and mandibular bones were involved (20%), though the occipital bone (13.3%) as well as the zygoma, maxilla, ethmoid, and calvarium (cumulatively accounting for 6.7%) were also affected [4]. A total of 20 reported cases of CMF have arisen in the sinonasal tract, but only four cases have been noted to involve the nasal septum [3].

Patient Demographic:

Sinonasal tract CMF is more likely to arise in women than men, with a 1.5:1 ratio, and is more common in patients over 40 years old [2]. Classically, CMF of the long bones was thought to be more common in young adults. However, CMF in the head/neck, including the sinonasal tract, seemingly has a predilection for females around the age of 60-80 years [5]. In particular, the documented cases of CMF involving the nasal septum have affected only female patients from ages 49-60 years old [3]. Our case report reflects these trends, though our female patient is the oldest (to date) to present with CMF of the nasal septum at the age of 71.

Symptoms:

With involvement of the sinonasal tract, presenting symptoms can include diplopia, pain, exophthalmos, neuralgia, epistaxis, nasal congestion/obstruction, headache, bony swelling, or chronic rhinosinusitis symptoms refractory to medical management [2,3]. However, it is frequently found incidentally in asymptomatic patients, as in the case of our patient [2]. Similarly, Wang et al described a patient presenting asymptomatically with a nasal septal CMF lesion found incidentally during work-up for congenital aural atresia [5].

Pathology

Grossly, CMF appears to be a firm but friable, fibrous, translucent, rubbery, yellow/gray/brown with a blue hue, and a peripherally lobulated mass [6]. Differentiating it from chondrosarcoma, CMF is distinctly demarcated from the surrounding bone [7]. CMF of the nasal septum, in particular, has been described as irregularly shaped, gray-white-tan in color with an elastic consistency [2,3]. Similarly, our patient's lesion was pink-tan, lobulated, and firm on gross examination.

Microscopically, CMF appears lobulated with both myxoid and chondroid matrix components and with concentrations of spindle and stellate cells along the fibroblastic periphery of lobules [8]. Compared to other CMF lesions, CMF of the skull/facial bones is most likely to have a microlobular pattern (versus a macrolobular pattern), calcification, permeation pattern of the bone, and liquefactive change [4]. CMF of

the skull/facial bones is least likely to have pleomorphic nuclei, cyst formation, giant cells, or necrosis [4]. Chondrosarcoma, on the other hand, has a uniform cellular arrangement of mostly chondroblasts and has a well-defined hyaline matrix, lacking a fibrous component [2,3]. Chondrosarcoma infiltrates and encases preexisting bony trabeculae [3,9]. Both CMF and chondrosarcoma may stain positively for vimentin, S-100, and SOX-9 transcription factor, making these unhelpful with diagnosis; a simple H&E stain remains the gold standard for pathological diagnosis [3,8].

In our case, the patient's pathology demonstrated a predominantly myxoid matrix with greater cellularity at the periphery of each lobule and with spindle and stellate cellular morphologies visualized throughout the majority of the lesion. No atypia or pleomorphism was noted and immunohistochemical stains were negative, including S-100. These findings support previous descriptions of CMF pathology specific to the skull/facial bones. Notably, about 22% of CMF cases are misdiagnosed and 11% of cases have no pre-surgical diagnosis [7]. When misdiagnosed as chondrosarcoma, dramatically aggressive surgical treatment, including amputation, has been erroneously pursued [6]. This underscores the importance of thorough microscopic review of the biopsy by pathologists in order to ensure appropriate surgical treatment.

Surgical Treatment

Jaffe et al originally advocated curettage and possible filling of the bony defect with bone chips or a bone graft [1].The lesion was thought to be benign, slow-growing, and unlikely to recur “even after mere curettage” [1]. However, recurrence up to 80% was noted with curettage only [7]. As more CMF cases have since been identified, the surgical treatment paradigm has shifted with most authors now advocating block resection [6,9].

In craniofacial CMF cases, complete surgical resection could result in significant morbidity and disfigurement. Reviewing nasal septal CMF cases, there is scant information on surgical treatment and approach, and there is no consensus on site-specific treatment of choice. Veras et al performed block resection only (their treatment of choice), thinking that curettage would increase recurrence risk [2]. Wang et al performed “complete removal” of a nasal septal lesion at the bony-cartilaginous junction via a trans-columellar approach [5]. Januszek et al surgically resected a nasal septal lesion with extension into the maxillay and sphenoid sinuses [10]. McClurg et al performed excision of a CMF lesion, nasal septum, left ethmoid, and left partial maxilla using a midfacial degloving via sublabial approach [3]. McClurg et al advocate complete surgical extirpation but state that curettage could be used for surgically inaccessible lesions, though repeat surgery and possible radiation therapy may be eventually required [3].

Similar to McClurg et al, we employed en bloc resection of the CMF lesion and it's origin (the nasal septum). Additionally, we also performed curettage of the cavity margins via a midfacial degloving approach. This approach was chosen over complete en bloc resection in order to minimize functional and aesthetic morbidity.

Surgical Complications

Potential complications of using a midfacial degloving approach are known to be nasal crusting, infranasal hypesthesia, nasal congestion, or epistaxis. McClurg et al did not note any of these complications in their patient [3]. Our patient did not have any of these complications related to the

surgical approach, though she did have persistent left epiphora for which dacryocystorhinostomy was performed. The epiphora may be attributable to post-operative scarring. In order to decrease the risk of this complication, Baujat et al placed lacrimal duct catheters during a paralateronasal approach for nasal bone CMF [8]. In the future, lacrimal duct catheterization with silicone tubing may beneficial during surgical treatment of patients with nasal septal CMF.

Recurrence and follow-up

The CMF recurrence rate is 11.5% [3]. The average age at recurrence is 22.6 years, typically occurring 5 months to 10 years after initial diagnosis, averaging at 38.4 months after initial diagnosis [4]. Risk of recurrence is thought to be higher in cases treated with curettage alone due to incomplete removal rather than regrowth [5]. One of the four documented cases of nasal septal CMF had recurrence after 12 months requiring re-operation [10]. Our patient had no recurrence noted on exam/MRI imaging at her 4-month follow-up. She will next be seen at a 12-month follow up visit with repeat MRI imaging.

Patients should be followed post-operatively to monitor for recurrence in addition to malignant transformation. CMF has a malignant transformation rate of 0.7%, and this risk is notably increased with the use of radiation therapy [3]. Due to this risk, multiple authors agree that radiation therapy should be reserved only for recurrences or primary tumors that are surgically inaccessible [3,7,8].

Conclusions

The presented case highlights the importance of including CMF in the differential diagnosis for patients with both symptomatic and asymptomatic nasal septal masses. Pathologic analysis, namely H&E stain, of the biopsy will facilitate differentiation from an aggressive chondrosarcoma and guide surgical planning. For CMF, particularly in the nasal septum, the currently accepted surgical management with block resection should be the goal, to minimize recurrence, but also be tailored to the patient, to minimize overall morbidity.

Figures

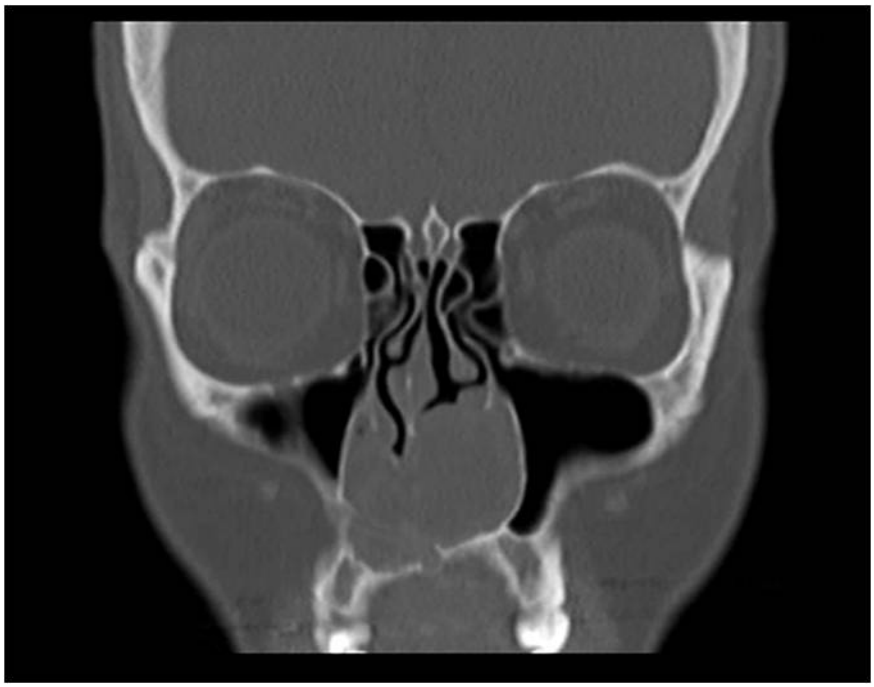

Figure 1: CT sinus of the patient, demonstrating a nasal septal lesion with locoregional mass effect and erosion of the hard palate.

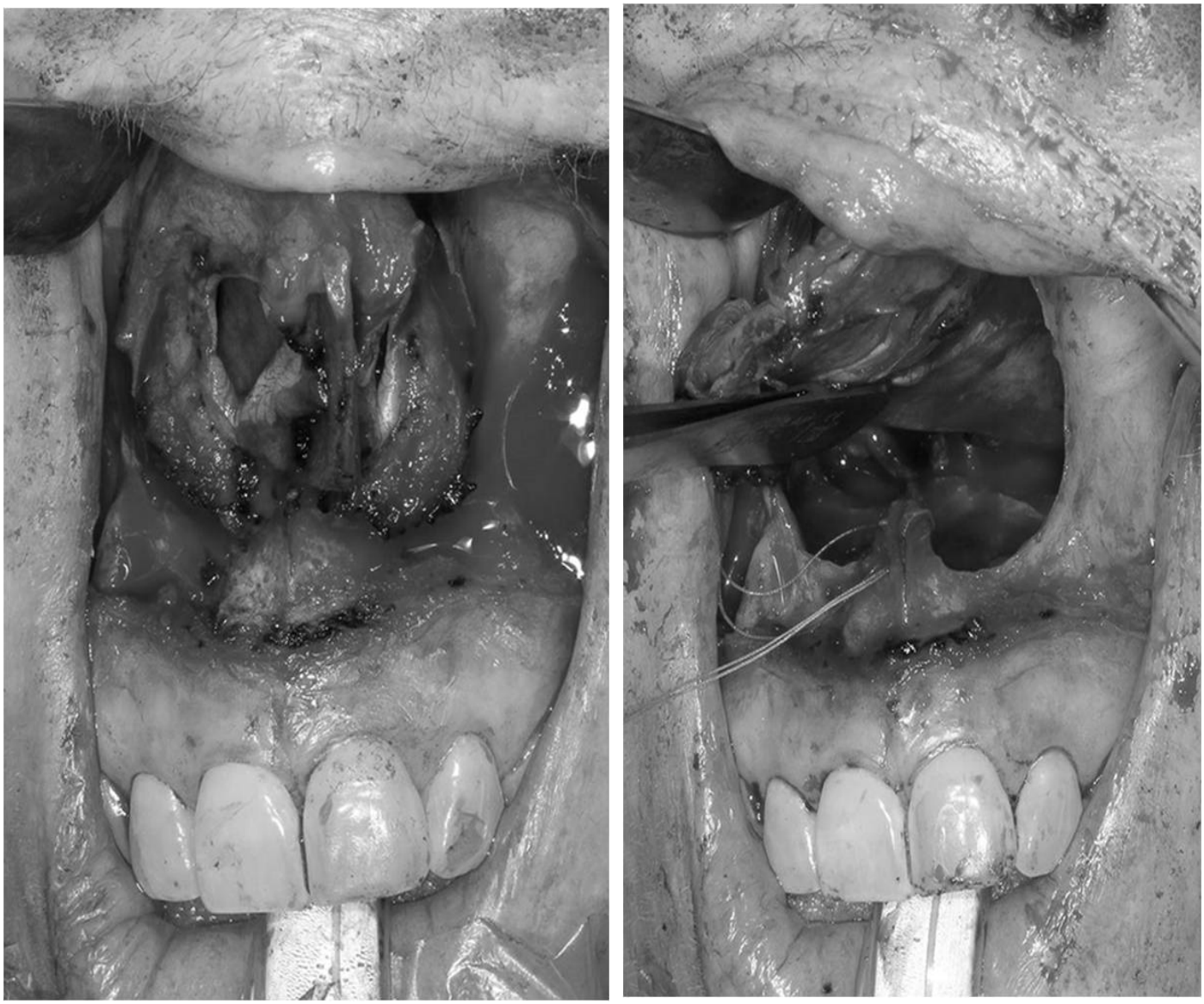

Figure 2: Intraoperative pre-excision (a) and post-excision (b) photographs of the nasal septal lesion.

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